TITLE: PERIOPERATIVE COMPLICATIONS ASSOCIATED WITH SPINAL FUSION IN RETT SYNDROME

Authors: Jenny Downs 1,2, Kingsley Wong 1, Peter Jacoby 1, Maree T. Izatt 3, Geoffrey N. Askin 3, Bruce McPhee 4, Peter Cundy 5,6, Helen Leonard 1

Organisation/ institution: 1 Telethon Kids Institute, The University of Western Australia, Perth, Australia; 2 School of Physiotherapy and Exercise Science, Curtin University, Perth, Australia; 3 Paediatric Spine Research Group, Queensland University of Technology and Mater Health Services, Brisbane, Australia; 4 Department of Surgery, University of Queensland, Brisbane, Australia; 5 Discipline of Orthopaedics and Trauma, University of Adelaide, Adelaide, Australia; 6 Department of Orthopaedic Surgery, Women's and Children's Hospital, Adelaide, Australia.

Introduction
Rett syndrome is a rare genetic neurodevelopmental disorder usually affecting females. Scoliosis occurs commonly and spinal fusion may be recommended if severe. We investigated short term complications of spinal fusion and the influence of pre-operative Cobb angle on aspects of the inpatient stay.

Methods
Data were ascertained from hospital medical records and the Australian Rett Syndrome Database, a longitudinal and population-based registry of Rett syndrome cases established in 1993. We calculated logistic regression models to assess the associations between Cobb angle (<70 vs >=70 degrees) and the development of a diagnosed respiratory complication, severe bleeding necessitating blood transfusion, ICU days (<2 vs >=2 days) and length of hospital stay (<14 vs >=14 days).

Results
Ninety-seven girls with Rett syndrome underwent spinal fusion at a median (range) age of 13 (7 to 18) years and hospital data were available for 87 (89.7%). The median (range) Cobb angle was 72 (41 to 126) degrees pre-operatively and 25 (4 to 61) post-operatively. A respiratory complication occurred in 35 (40.2%) and severe bleeding in 32 (36.4%). Over-sedation (8.0%), wound infection (2.3%), spinal cord injury (2.3%) and cardiac arrest (1.1%) occurred infrequently. Taking into account the effects of age of scoliosis onset, the pre-operative Cobb angle was not associated with the presence of respiratory complications or severe bleeding. Girls with a pre-operative Cobb angle >=70 degrees had more than twice the odds of a longer ICU stay (OR 2.61, 95%CI 0.94, 7.26) but their overall length of hospital stay was not greater (OR 0.89, 95%CI 0.33, 2.43).

Conclusion
Respiratory complications and severe bleeding occurred commonly following spinal fusion, but the extent of respiratory complications was greater in those with a larger pre-operative curve. Earlier spinal fusion has potential to reduce the short term complications of spinal fusion and, as a consequence, reduce hospital costs.

Word count: 300 (max. 300 words)